Meigs' and Pseudo-Meigs' syndrome

Abstract
Diagnosing Meigs' syndrome is challenging in that it can be mistaken for a number of other conditions. Ultrasound can identify ascites and pleural effusions, which is essential in accurate identification.

Keywords: Meigs, ovarian tumours, ultrasound.

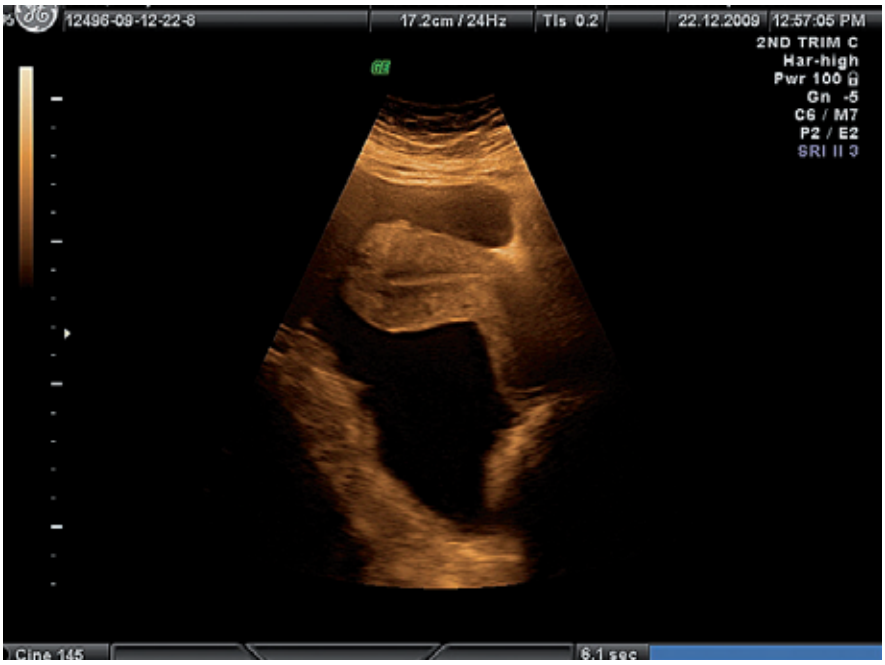

Figure 1: Ascites.

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Introduction

Meigs' syndrome is diagnosed based on a triad of an ovarian fibroma, pleural effusion and ascites.It resolves spontaneously after the resection of the fibroma.[1] In 1852, Blin published the description of an ovarian fibroma with abdominal effusion in the Société de Biology de Paris (cited by Lallemand).[2] A Demons of Bordeaux, France, gave a report to the Société de Chirurgie de Paris in 1887, that nine of 50 patients with ovarian cysts were cured of their ascites and hydrothorax by removal of the adnexal cyst. In 1937, Joe Vincent Meigs (1892–1963), an American professor of the Harvard Medical School of Gynaecology drew widespread attention of the medical profession to the syndrome.[3] Meigs used seven cases to highlight the association between a fibroma of the ovary, ascites, and hydrothorax. It was coined as Meigs' syndrome in 1937 by Rhodes and Terrell.[4]

The following four characteristics were selected by Meigs in 1945 to define the syndrome:

- The tumour is a benign fibroma or a fibroma-like tumour of the ovary (such as thecoma and granulosa cell tumours)
- Ascites
- Pleural effusion(s)
- Removal of tumour must cure the patient.[5]

Other benign cysts of the ovary (such as struma ovarii, mucinous cystadenoma and teratomas), leiomyoma of the uterus, and secondary metastatic tumours to ovary if associated with hydrothorax are referred to as 'Pseudo-Meigs" syndrome.[6]

An atypical case of Meigs' syndrome was reported in 1990 by Martin, *et al.*[7] presenting as bilateral sanguineous pleural effusion without ascites in a woman with a granulosa cell tumour.

Epidemiology

Meigs' syndrome accounts for about 1% of ovarian tumours, and ovarian fibromata are found in 2–5% of surgically removed ovarian tumours. About 10–15% of women with an ovarian fibroma have ascites, and 1% has hydrothorax. Approximately 70% of pleural effusions are right-sided, 15% left sided, and 15% are bilateral.[8,9] With appropriate management, life expectancy after surgical removal of the tumour is same as the general population. Meigs' syndrome is very uncommon before the third decade but progressively increases thereafter to peak in the seventh decade.

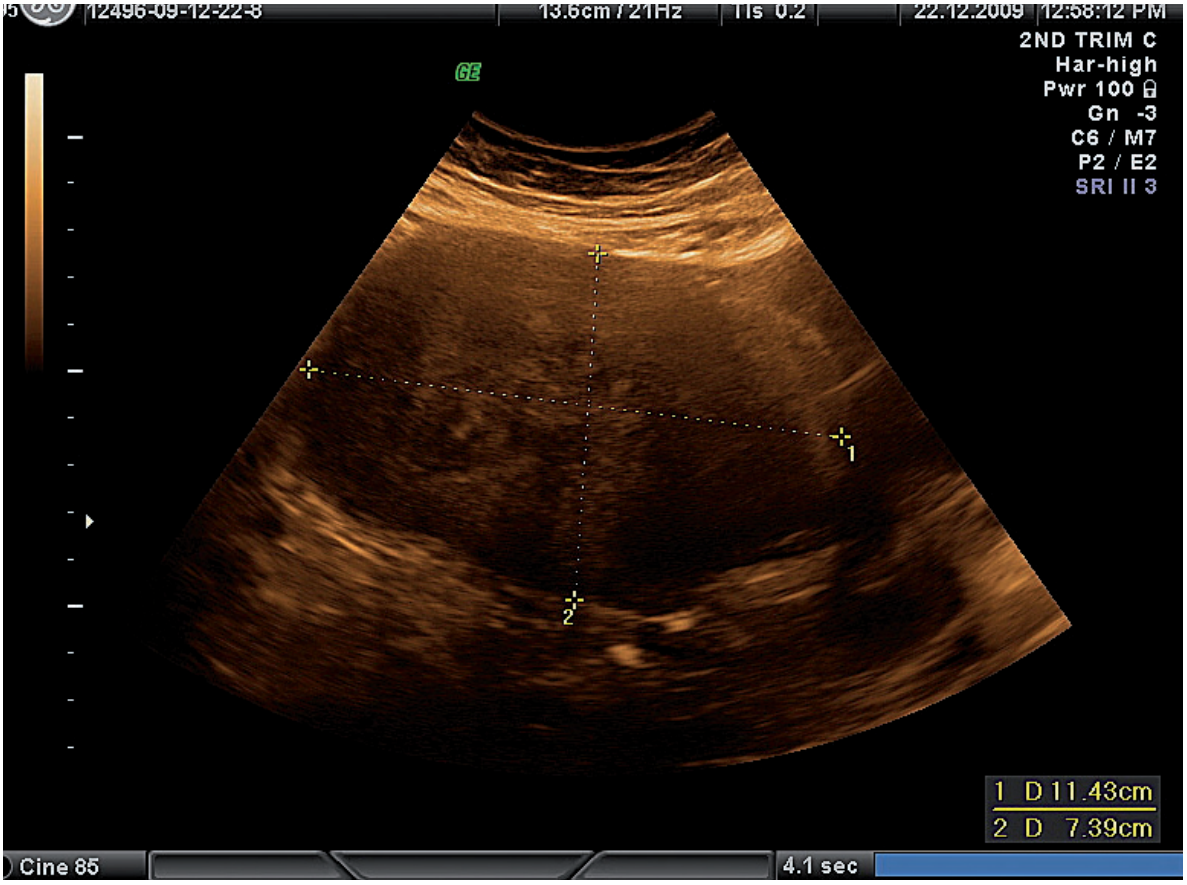

Figure 2: Solid right sided ovarian mass.

Pathophysiology

The exact pathogenesis of the ascites is still unknown. A possible theory is that there is filtration of interstitial liquid into the peritoneum through the ovarian tumour capsule. This then moves from the peritoneal cavity to the pleural cavity through diaphragmatic defects or via the lymphatic channels and eventually causes an exudative pleural effusion.[5,9] An imbalance between the blood supply to a large tumour and its venous and lymphatic drainage may be responsible for stromal oedema and transudation.[10] However, some new studies suggest the fluid accumulation may be related to proteins such as vascular endothelial growth factor (VEGF) that raise capillary permeability.[11]

There are some published case reports of Meigs' syndrome with elevated CA 125 levels.[12] The precise mechanism is not clearly known, however, immunohistochemical staining for CA 125 suggests that serum elevation of CA 125 antigen is secondary to mesothelial expression of CA 125.[13] Biochemical factors, mechanical irritation from a large tumour,and a raised intraperitoneal pressure secondary to ascites are possible primary factors.

Case report

A 66-year-old female was referred to a tertiary care facility for diagnostic work-up of a known pleural effusion, ascites and a significantly raised Ca125 (759 kU/L). The pleural effusion was diagnosed six months prior to presentation and yielded negative cytology on aspiration. The patient re-presented with marked abdominal discomfort and bloating. An abdominopelvic ultrasound revealed significant ascites (Figure 1). In addition, an 11cm adnexal mass, with sonographic appearance consistent with a solid ovarian mass (suggestive of an ovarian fibroma) was visualised (Figure 2). Abdominal paracentesis revealed no evidence of malignant cells and the provisional diagnosis of Meigs' syndrome was considered. The patient underwent explorative laparotomy followed by a total abdominal hysterectomy and bilateral salpingooophorectomy. Her postoperative course was uneventful and she made a complete recovery. Pathology confirmed a right sided ovarian fibroma (Figure 3).

Discussion

The combination of ascites and raised Ca125 in an elderly lady raises the suspicion about potential ovarian malignancy. Meigs' syndrome however, is a clinical and surgical syndrome that tests our diagnostic abilities and it is an important diagnosis from a patient's perspective.

The differential diagnosis for the presenting signs and symptoms include malignant ovarian tumour, other cancers including bowel and lung, nephrotic syndrome, congestive cardiac failure, liver cirrhosis and tuberculosis.

In spite of the valuable contribution of medical imaging techniques, the presumptive diagnosis of Meigs' syndrome is made clinically. Upper abdominal ultrasound demonstrates ascites and should detect pleural effusions. Pelvic ultrasound demonstrates the presence of a well demarcated adnexal mass without increased vascularity. However, fibromas and fibrothecomas

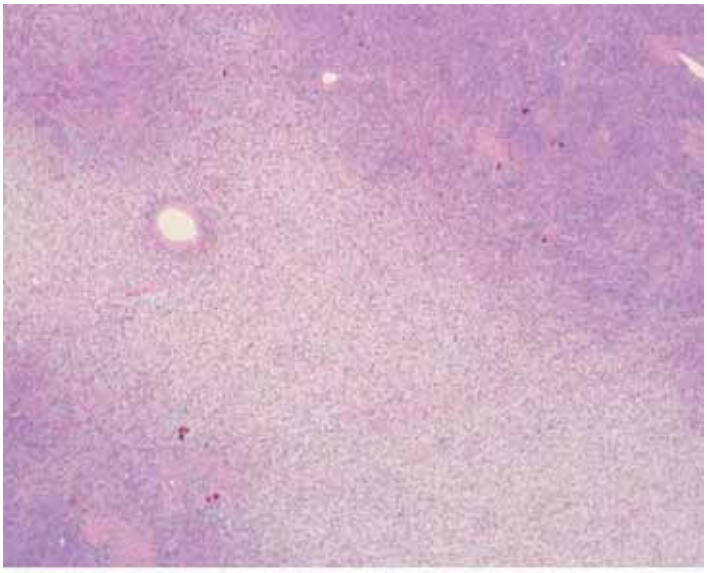

Figure 3: Histopathology confirming ovarian fibroma.

can display a wide variety of sonographic features,[14] particularly when changes such as oedema, degeneration, haemorrhage, and are present. Chest x-ray may be used to confirm the presence of a pleural effusion. Other imaging modalities like MRI or CT can be considered to exclude metastatic disease prior to treatment.

The definitive diagnosis of Meigs' syndrome is usually postoperative with resolution of ascites and pleural effusions, and histological confirmation of the tumour.

Medical care of patient with Meigs' syndrome involves paracentesis and thoracentesis for ascites and pleural effusion respectively. The treatment of choice is exploratory laparotomy with surgery and staging. Frozen section of ovarian mass is performed to confirm the benign nature of the mass. In women of reproductive age, unilateral salpingo-oophorectomy is the treatment of choice, whereas in post-menopausal women treatment is total abdominal hysterectomy with bilateral salpingo-oophorectomy.

Conclusion

An awareness of benign lesions of the pelvis with associated adverse features is important for both clinicians and their imaging partners to limited patient anxiety and direct appropriate treatment. Biochemical markers like Ca125 have limited diagnostic capabilities and their real value lies in cancer treatment surveillance.

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